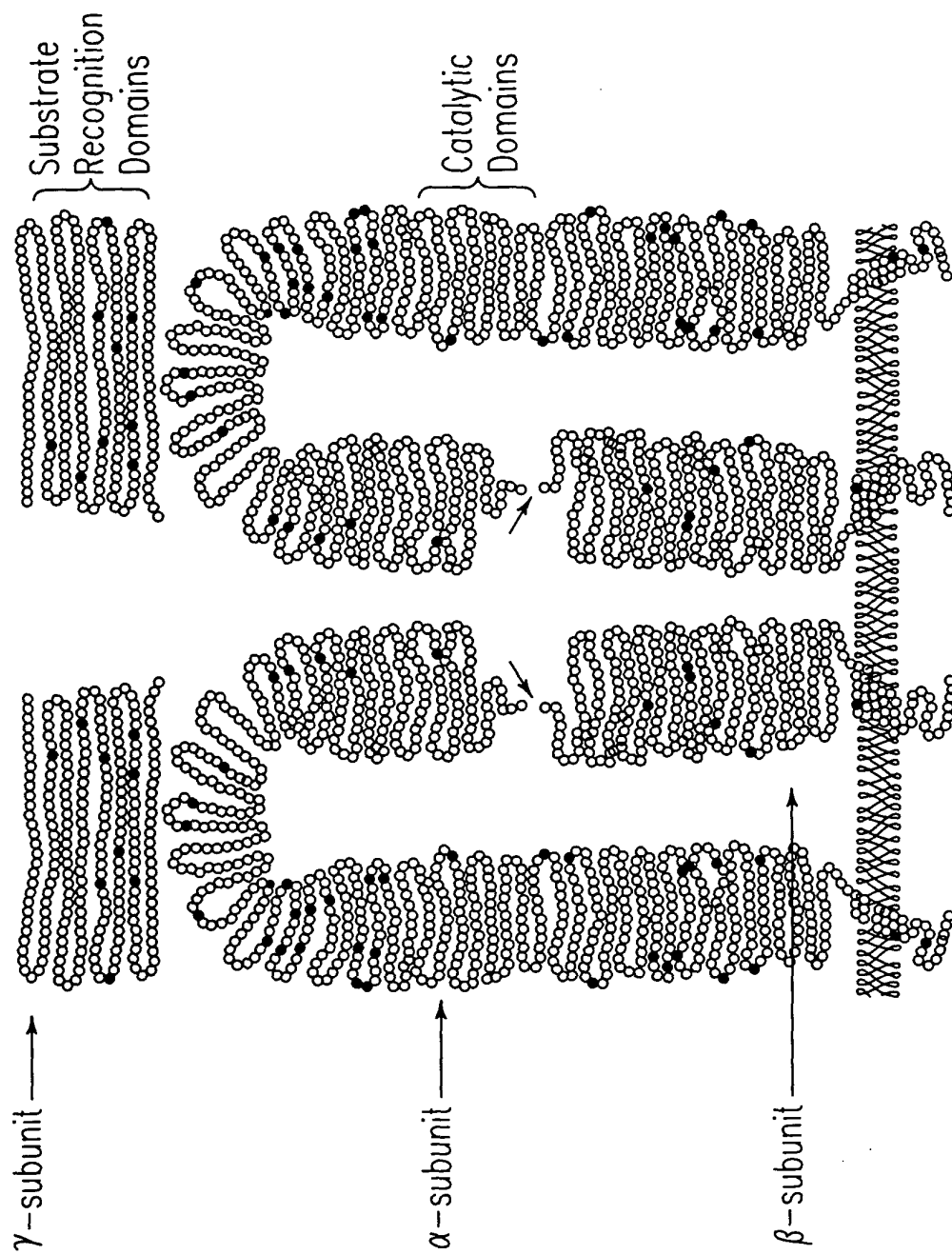


FIG. 1



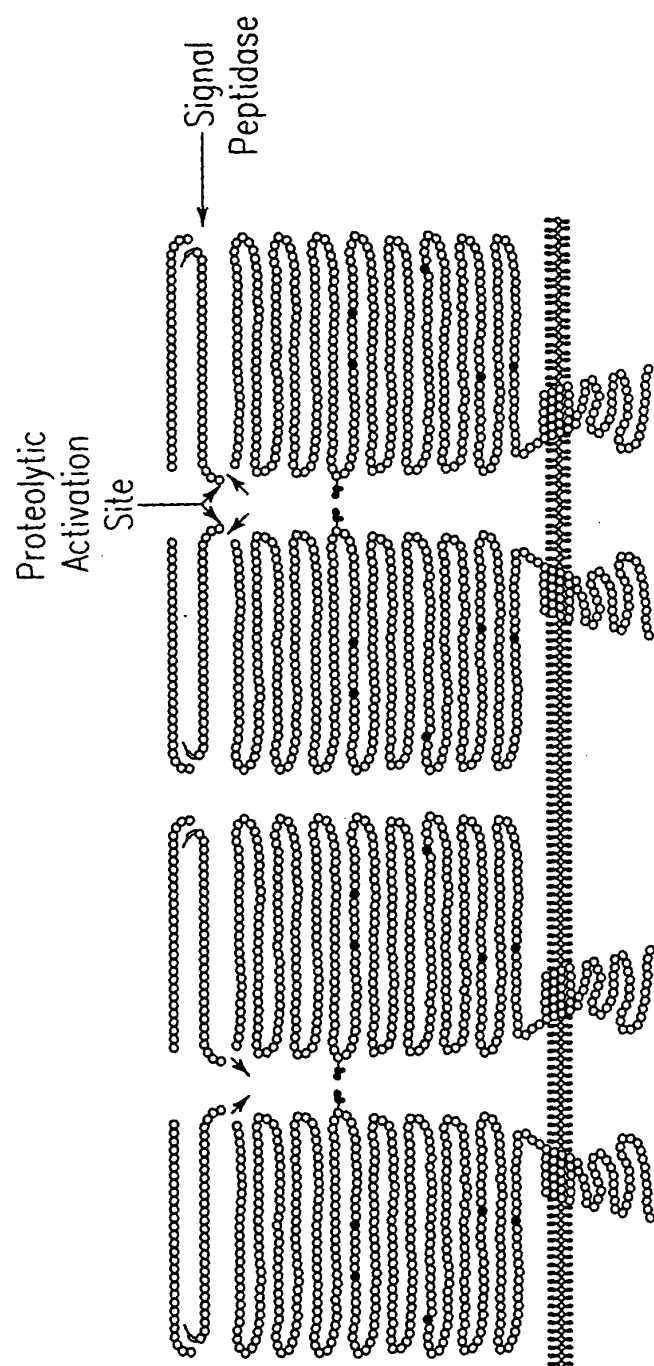


FIG. 2

The diagram illustrates three pathways for the degradation of N-linked glycoproteins, showing the progression from the Golgi apparatus to the extracellular space or lysosomes.

- Pathway 1 (Normal):** An N-linked glycoprotein with a core fucose (triangle) and mannose residues (squares) is processed by Glc II and Glc I in the ER. It then moves through the Golgi (Gn I, Gn II, Gn III) and is phosphorylated by GnPT to form a mannose 6-phosphate (M6P) residue. This M6P-tagged protein is then transported to the lysosome via M6P receptors for degradation by Lysosomal Phosphatases & Mannosidases.
- Pathway 2 (Mannose 6-phosphate pathway):** Similar to Pathway 1, but the protein is not phosphorylated by GnPT. Instead, it is directly transported to the lysosome via M6P receptors. In the lysosome, it is degraded by Lysosomal Phosphatases & Mannosidases.
- Pathway 3 (Deoxymannosidosis):** The protein is not phosphorylated by GnPT and is instead transported to the extracellular space. In the extracellular space, it is degraded by UCE (Extracellular Uridylate Carboxylase) and GnPT (Golgi Nucleoside Phosphorylase) to form a deoxymannoside derivative.

←

Acid α -glucosidase

Pathway 2

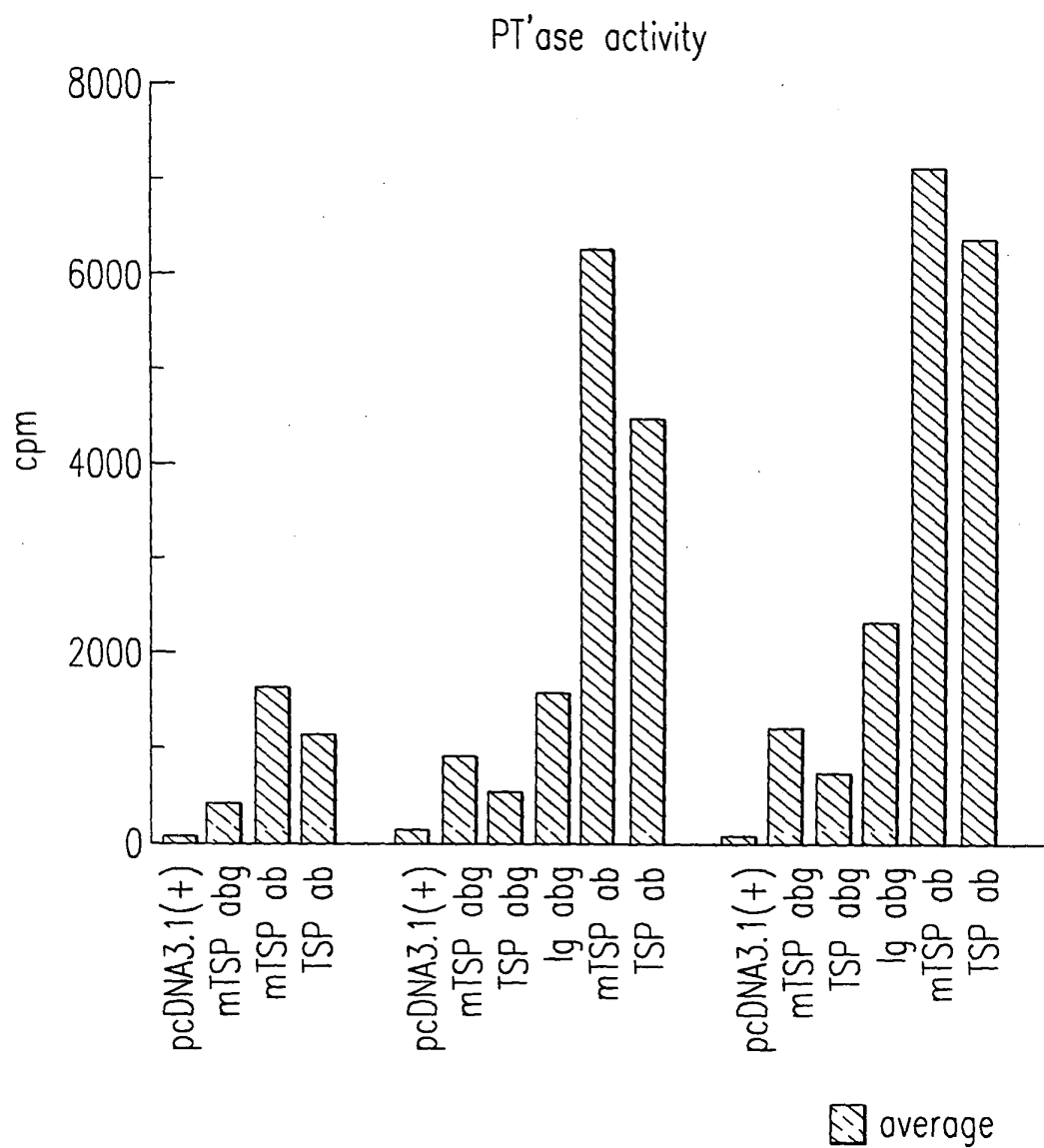


FIG. 4